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Myelodysplastic Syndromes - MDS - Subtypes and Classification [1]

This section has been reviewed and approved by the Cancer.Net Editorial Board [2], 10/2014

ON THIS PAGE: You will learn about how doctors describe MDS. This is called subtype and classification. To see other pages, use the menu on the side of your screen.

MDS is classified into several different subtypes, depending on blood cell counts, the percentage of blasts in the bone marrow, and the risk that it will turn into AML. Also, MDS is classified as either primary MDS or secondary MDS, and it may be given a disease staging called an IPSS-R score. These classifications help doctors plan treatment and predict a patient?s prognosis, which is the chance of recovery. Each is described below in more detail.

WHO system for MDS subtypes

The World Health Organization (WHO) developed a classification system for MDS to standardize the definitions of the different subtypes. The seven subtypes of MDS in this system include:

Refractory anemia (RA). The primary sign of RA is anemia. White blood cell counts and platelet counts are normal. There are less than 5% blasts found in the bone marrow. This subtype of MDS does not often turn into AML.

Refractory anemia with ringed sideroblasts (RARS). People with this subtype of MDS have anemia, similar to those with RA, except more than 15% of the red blood cells are sideroblasts. A sideroblast is a red blood cell in which the iron in the cell appears to be in a ring around the center of the cell where the genes are found, called the nucleus. The white blood cell and platelet cell counts are usually normal. People diagnosed with RARS have a low risk of developing AML.

Refractory cytopenia with multilineage dysplasia (RCMD). In this subtype, people have less than 5% blasts and less than 15% ringed sideroblasts in the bone marrow. The other bone marrow cells look abnormal when viewed under the microscope. At least two of the blood cell counts are low. RCMD may eventually turn into AML.

Refractory cytopenia with multilineage dysplasia and ringed sideroblasts (RCMD-RS). This subtype is similar to RARS, in which people have anemia and more than 15% sideroblasts. The

other bone marrow cells also look abnormal when viewed with a microscope. In addition, at least two types of blood cell counts are low. RCMD-RS may eventually turn into AML.

Refractory anemia with excess blasts (RAEB). People with RAEB can have decreases in all or some of their blood cell counts. There are less than 5% blast cells in the blood and 5% to 20% blasts in the bone marrow. People with more than 20% blasts in the bone marrow are diagnosed with AML. People with RAEB may also have lower white blood cell and platelet counts. About 40% of people diagnosed with RAEB eventually develop AML.

Myelodysplastic syndrome, unclassified (MDS-U). People diagnosed with this subtype have decreased numbers of white blood cells, red blood cells, or platelets, but do not have the specific signs of the other MDS subtypes.

MDS associated with isolated del(5q). People with this subtype have anemia and fewer than 5% blasts, and genetic material is missing from chromosome 5.

CMML and JMML. In addition to the seven MDS subtypes above, chronic myelomonocytic leukemia (CMML) and juvenile myelomonocytic leukemia (JMML) are types of blood cancers that the WHO classifies as ?mixed myelodysplastic/myeloproliferative diseases.? Unlike other types of MDS in which blood counts are low, white blood cell counts are higher in these subtypes. Both CMML and JMML begin after a change, or mutation, happens in a type of blood cell called a monocyte. CMML generally occurs in people ages 65 to 75. JMML is most common in children younger than 6. Treatment is similar to MDS and can include chemotherapy and/or stem cell transplantation (see Treatment Options [3]).

Primary/secondary MDS

In addition to subtype, MDS is called either primary or secondary MDS. Primary MDS is much more common than secondary MDS. About 80% of people with MDS have primary MDS.

- In primary MDS, no apparent risk factors can be found. This may also be called de novo MDS.
- Secondary MDS occurs because of damage to the DNA from chemotherapy or radiation therapy previously given to treat another medical condition. MDS can develop two to 10 years after such treatment. Secondary MDS is often associated with more complex chromosomal abnormalities.

IPSS-R system

The revised International Prognostic Scoring System (IPSS-R) is another classification system used by doctors to help predict a person?s risk of developing AML and overall survival. The IPSS-R looks at factors such as the percentage of blasts found in the bone marrow, type and extent of chromosomal changes, and levels of hemoglobin found in red blood cells, platelets, and a type of white blood cell called neutrophils.

Poor prognostic factors include:

- Specific or many chromosomal changes

- Many blasts in the bone marrow
- Low levels of hemoglobin, platelets, and neutrophils

The total IPSS-R score places people with MDS into five distinct groups: very low risk, low risk, intermediate risk, high risk, and very high risk. People with MDS who have a lower IPSS-R score have the best outlook for survival and need less aggressive treatment. For patients with lower IPSS-R scores, overall survival rates tend to be lower when they need red blood cell transfusions, a procedure in which blood or blood cells from one person are given to another person, compared to those who do not need transfusions. A person diagnosed with a high-risk subtype of MDS and whose IPSS-R score is high usually needs more intensive treatment.

Recurrent: Recurrent MDS is MDS that has come back after a period of remission, or absence of symptoms, also called ?no evidence of disease? or NED. If there is a recurrence, the subtype and IPSS-R score may need to be determined again using the systems above.

Information about the MDS subtype and classification will help the doctor recommend a treatment plan for you. The next section helps explain the treatment options for MDS. Use the menu on the side of your screen to select Treatment Options, or you can select another section, to continue reading this guide.

Links:
[1] http://www.cancer.net/cancer-types/myelodysplastic-syndromes-mds/subtypes-and-classification
[2] http://www.cancer.net/about-us
[3] http://www.cancer.net/node/19387